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## A rare presentation of midgut malrotation as an acute intestinal obstruction in an adult: Two case reports and literature review

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## ABSTRACT

**INTRODUCTION:** Midgut malrotation is a congenital anomaly presenting mainly in the childhood. Its presentation as an acute intestinal obstruction is extremely rare in adults usually recognized intra-operatively, therefore a high index of suspicion is always required when dealing with any case of acute intestinal obstruction.

**PRESENTATION OF CASE:** We report two cases of young adults who presented with symptoms of acute intestinal obstruction and were diagnosed intra-operatively as cecal volvulus and paraduodenal hernia, respectively, caused by midgut malrotation. Post-operative CT scan confirmed these findings.

**DISCUSSION:** Malrotation of the intestinal tract is a product of an aberrant embryology. The presentation of intestinal malrotation in adults is rare (0.2–0.5%). Contrast enhanced CT can show the abnormal anatomic location of a right sided small bowel, a left-sided colon and an abnormal relationship of the superior mesenteric vein (SMV) situated to the left of the superior mesenteric artery (SMA) instead of to the right.

**CONCLUSION:** Anomalies like midgut malrotation can present as an operative surprise and awareness regarding these anomalies can help surgeons deal with these conditions.

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## 1. Introduction

Midgut malrotation is a congenital anomaly referring to either lack of or incomplete rotation of the fetal intestines around the axis of the superior mesenteric artery during fetal development. Most patients usually present with bilious vomiting in the first month of life because of duodenal obstruction or a volvulus. It is rare for this condition to present in adulthood. It has been estimated that it affects approximately 1 in 500 live births.<sup>1</sup> More than 90% of patients will present by the time of their first birthday.<sup>2</sup> It has been reported that the incidence of malrotation in adults is approximately between 0.0001% and 0.19%.<sup>2,3</sup> True incidence is very difficult to measure, as many cases remain asymptomatic. In adults, the diagnosis in the pre-operative period is extremely difficult owing to a very low index of suspicion.

We report two cases of acute intestinal obstruction in adults with midgut malrotation as the cause.

## 2. Case report 1

A 26-year-old male patient was admitted in the emergency ward of Safdarjang Hospital with complaints of generalized pain

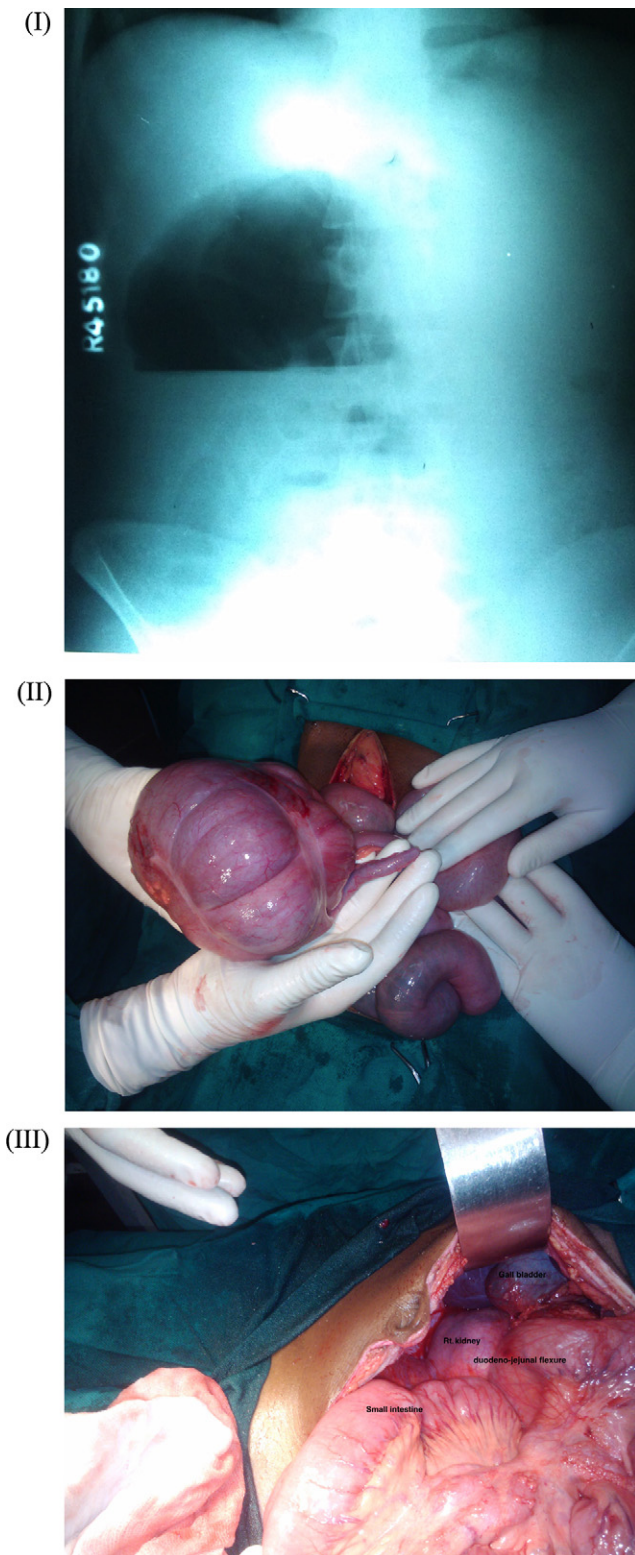
in abdomen for 3 days. The pain was colicky in nature. The patient also vomited thrice on the day of presentation with vomitus containing biliary contents. He was also complaining of severe anorexia and inability to pass stools and flatus for 2 days. There was no history of similar complaints in the past and he also denied any history of tuberculosis, peptic ulcer disease or any other chronic medical illness. He had no history of any abdominal surgery (Figs. 1 and 2).

On physical examination, the patient was a well-nourished young adult male. The patient was having tachycardia with 110-pulse rate and 104/60 mm Hg of B.P. The abdomen was distended with increased bowel sounds on auscultation. Per rectal examination did not reveal any abnormality. The patient's full blood count demonstrated a hemoglobin level of 14.1 g/dl and mild leukocytosis of 13,600/mm<sup>3</sup> with prominent neutrophilia. All other biochemical parameters including blood sugar, serum electrolytes, liver function tests, kidney function tests, clotting profile, amylase and lipase levels were within normal limit. Abdominal X-ray revealed a large dilated bowel loop on the right side with suspicion of a cecal volvulus. A diagnosis of acute intestinal obstruction was made and the patient was admitted and planned for urgent exploratory laparotomy.

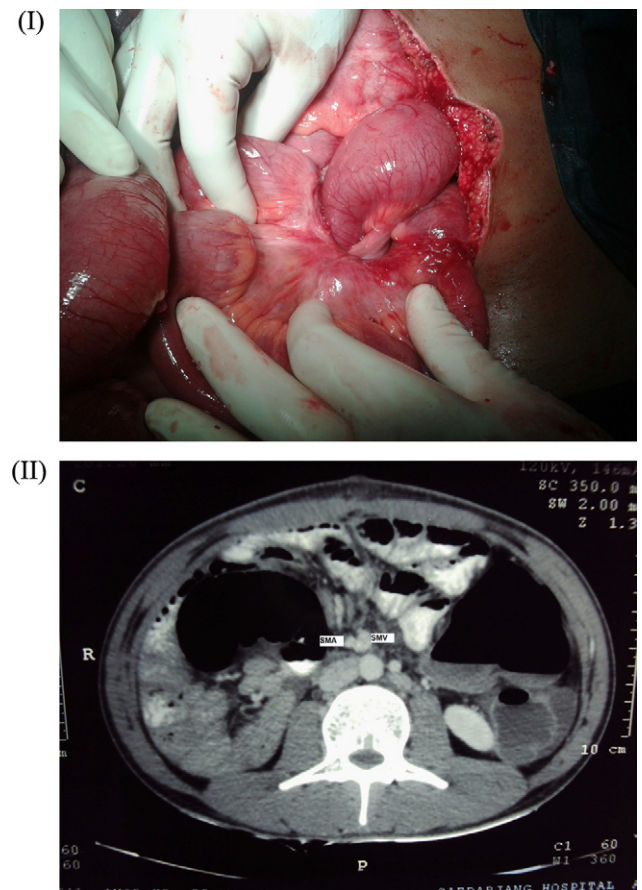
The abdomen was opened from a midline incision and a hugely dilated loop of cecum with appendix and ascending colon visualized. The whole small bowel loops were seen to be mainly concentrated on the right upper abdomen with duodenojejunal junction present abnormally on the right side of midline and there was inability to visualize the hepatic flexure and transverse colon. On careful inspection a large cecal volvulus noted involving cecum, appendix and ascending colon which also showed a few serosal

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**Fig. 1.** (1) X-ray showing cecal volvulus. (2) Cecal volvulus showing dilated cecum with appendix. (3) Right side of peritoneal cavity showing absence of ascending colon, hepatic flexure, and transverse colon. Doudeno-jejunal flexure can be seen abnormally located on the right side. Kidney can also be visualized.



**Fig. 2.** (1) A paraduodenal hernia. A large defect with a loop of small intestine trapped in it can be seen. (2) ACT scan showing the abnormal relationship of superior mesenteric vein (SMV) with the superior mesenteric artery (SMA).

tears and the distal end, i.e. transverse colon, going behind the peritoneum in the right iliac fossa. The peritoneum was then carefully incised to trace the transverse colon, which was going directly from the right iliac fossa to the left hypochondrium to continue as splenic flexure. After full mobilization of the involved bowel right hemicolectomy was done and intestinal continuity was maintained with a single layer end to end ileocolic anastomosis. Post-operative period was uneventful and the patient was discharged on the 6th post-op day. The patient is on a regular follow up. Post-operative CT scan of the abdomen showed alteration in the relation of the superior mesenteric artery with the vein.

### 3. Case report 2

A 20-year-old male patient was admitted in the emergency ward with the complaints of generalized pain in abdomen, which was colicky in nature. The patient also had multiple episodes of vomiting for 6 days, which was bilious. It was associated with fever and inability to pass stools and flatus for 2 days. The patient was nonalcoholic with no history of any trauma, chronic major illness, previous surgery or any drug intake.

On examination the patient was having tachycardia with 120/min pulse rate and 96/60 mm Hg of blood pressure. On per abdominal examination mild distension was present with diminished bowel sounds. Per rectal examination did not reveal any abnormality. Blood drawn for investigations and nasogastric tube and urinary catheter was inserted.



Routine blood investigations including blood sugar, serum electrolytes, liver function test, kidney function test, serum amylase and arterial blood gases report were within normal limits. Full blood counts revealed hemoglobin level of 13.6 g/dl and total leukocyte count of 17,200/mm<sup>3</sup> with predominantly neutrophilia. X-ray abdomen revealed absence of gas under diaphragm and multiple air fluid levels. USG scan showed dilated small intestinal loops with normal liver, spleen and pancreas.

Diagnosis of acute intestinal obstruction was made and the patient was taken up for exploratory laparotomy. Intra-operatively internal herniation of the small bowel entrapped in the paraduodenal recess was seen. Constricting neck of the hernia was relieved and the entrapped small intestine was relieved to see two small intestinal strictures and multiple impending perforations at the site of constriction ring. The whole proximal small intestine was seen to be edematous and dilated. Strictured segment of small intestine resected and a double barrel ileostomy was made because of the poor general condition and severe edema and dilatation of proximal small intestine and abdomen was left open with Bogota bag placement to prevent abdominal compartment syndrome. In the postoperative period the patient was comfortable with no obvious complications.

Postoperative CT scan showed an abnormal course of transverse colon, which is partly retroperitoneal and going behind the third part of the duodenum.

Ileostomy started functioning on 2nd day and patient was taken up for delayed abdominal closure on 4th postoperative day. After 3 months ileostomy was closed. Postoperative period was uneventful and the patient is on a regular follow up.

#### 4. Discussion

The cecum is the proximal portion of the colon after the ileocecal valve. It measures 6 cm in length and 7.5–8 cm in width. The cecum is generally covered by peritoneum, although in most cases, there is no distinct mesentery and the mobility is limited. Occasionally, the cecum can be particularly mobile, which predisposes to cecal volvulus and may contribute to unusual clinical presentations of acute appendicitis.<sup>7,8</sup>

The cecal volvulus is usually presented as distal small bowel obstruction. Plain films of the abdomen show distal small bowel obstruction with a dilated cecum in the epigastrium or the left upper quadrant. Barium enema is usually diagnostic, showing the level of obstruction and the ileocecal junction to the right of the cecal bubble. The goals of surgery are to decompress the obstruction and fix the cecum in the right lower quadrant, either by creating a pocket in the parietal peritoneum and suturing the cecum to the peritoneum or by using tube cecostomy. If strangulation has occurred, right hemicolectomy is the definitive procedure.

In a study published by Fukuya et al., seven adult patients presented with midgut volvulus as a result of intestinal malrotation. Four patients presented with long histories of intermittent abdominal pain, three patients presented with acute onset of severe abdominal pain. Abdominal angiography in one of these patients showed abnormal courses of mesenteric vessels to the volvulized segment of small bowel. These three patients with the acute onset had to undergo laparotomy, which showed ischemic segments of bowel.<sup>4</sup>

Malrotation of the intestinal tract is a product of a well-defined aberrant embryology. Because the consequences of malrotation associated with midgut volvulus may be catastrophic, an understanding of the anatomy, diagnostic criteria and appropriate therapy for this emergency condition is imperative.<sup>5</sup> The presentation of intestinal malrotation in adults is rare, and occurs in approximately 0.2–0.5%. Contrast enhanced CT can show the abnormal

anatomic location of a right sided small bowel, a left-sided colon and an abnormal relationship of the superior mesenteric vein (SMV) situated to the left of the superior mesenteric artery (SMA) instead of to the right and the characteristic 'whirlpool or whirl sign' describing the swirling appearance of bowel and mesentery twisted around the superior mesenteric arterial axis with the presence of midgut volvulus.<sup>6</sup>

Congenital internal hernias are the result of malformation of the peritoneum and, in some instances, midgut malrotation during the embryonic period. Internal hernias, either congenital or acquired, account for a small percentage of small bowel obstruction cases (0.6–5.8%).<sup>9–13</sup> This condition involves herniation of a viscus, usually small bowel, through a normal or abnormal aperture within the peritoneal cavity. This herniation may be intermittent or persistent and may pose a diagnostic challenge given the rare nature of its occurrence. Because of the risk for strangulation of the hernia contents, even small internal hernias are dangerous and may be lethal. However, as a result of their rarity, discovery of an internal hernia at laparotomy may be confusing to an unsuspecting surgeon who is not familiar with this abnormality, and thus appropriate management may be compromised. A paraduodenal hernia (PDH) is the most common variety of congenital internal hernia, followed by the transmesenteric and transomental varieties.

Also known as a paramesocolic hernia, PDH accounts for nearly 53% of the 500 published series of all internal hernias and were first described by Neubaur in 1786.<sup>14–18</sup> One hundred years later, Treitz and Waldeyer additionally described *hernia retroperitonealis*—several peritoneal folds and fossae through which small bowel could potentially herniate. The pathogenesis of PDH is controversial, but two theories regarding its origin appear to be most popular.<sup>19</sup> In a report by Moynihan in 1889, it was proposed that paraduodenal fossae were congenital and the hernia was acquired by gradual enlargement of an existing fossa. In 1923, Andrews disputed this theory and proposed that PDH forms as a result of a congenital anomaly in development of the peritoneum that arises during midgut rotation. The average age at diagnosis is reported to be between the third and fourth decades of life. Approximately 75% occur on the left at the paraduodenal fossa of Landzert. Right paraduodenal hernias occur through an abnormal pocket of jejunal mesentery next to the duodenum and immediately behind the superior mesenteric artery.<sup>20</sup>

A PDH originates from abnormalities arising during the second phase of embryonic intestinal rotation; it results in arrest of further rotation of the pre-arterial segment of the gut in the right side of the abdomen. Continued rotation of the postarterial segment leads to entrapment of small bowel behind the right colonic mesentery, with the superior mesenteric artery forming the anterior edge of the hernia sac.<sup>21</sup>

PDH may be asymptomatic and be discovered incidentally at laparotomy, at autopsy, or during radiologic studies for other unrelated causes.<sup>19,22,23</sup> More commonly, PDH is manifested as acute small bowel obstruction on a background of recurrent chronic, vague abdominal pain. The abdominal pain associated with a left PDH is typically left sided but can be variable in location and sometimes even right sided.<sup>24</sup>

Treatment of PDH follows the basic principles of hernia surgery—reduction of the contents, resection of the hernia sac, restoration of normal bowel anatomy, and repair of the hernia defect. The small bowel may be manually reduced if the hernia orifice is large enough and the defect can be closed with non-absorbable suture.<sup>19,25</sup>

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## Ethical approval

Informed consent from the patient obtained.

## Author contributions

SS performed the operations, involved in the pre-operative work up and post-operative care, conceived the write up, and performed the literature search and manuscript preparation.

AD assisted during the operations and was involved in the pre-operative work-up and post-operative care of the patient.

ASC and SVA helped in the preparation of the manuscript and were involved in the pre-operative work-up and post-operative care of the patient.

JC assisted during the operations.

All authors read and approved the manuscript for submission.

## References

- Torres AM, Ziegler MM. Malrotation of the intestine. *World Journal of Surgery* 1993;**17**:326–31.
- Von Flue M, Herzog U, Ackermann C, Tondelli P, Harder F. Acute and chronic presentation of intestinal non-rotation in adult. *Diseases of the Colon and Rectum* 1994;**37**:192–8.
- Wang C, Welch C. Anomalies of intestinal rotation in adolescents and adults. *Surgery* 1963;**54**:839–55.
- Fukuya T, Brown BP, Lu CC. Midgut volvulus as a complication of intestinal malrotation in adults. *Digestive Diseases and Sciences* 1993;**38**(3):438–44.
- Margarita Torres A, Ziegler MM. Malrotation of the intestine. *World Journal of Surgery* 1993;**17**:326–31.
- Sahu SK, Raghuvanshi S, Sanchan PK. Adult malrotation presenting as midgut volvulus: case report. *Journal of Surgical Arts* 2012;**5**.
- Pinto A, Di Raimondo D, Tuttolomondo A, Fernandez P, Caronia A, Lagalla R, et al. An atypical clinical presentation of acute appendicitis in a young man with midgut malrotation. *Radiography* 2007;**13**(2):164–8.
- Luca S, Pemberton JH. Embryology and anatomy of the colon. *Shackelford's surgery of the alimentary tract*, vol. 2(4), 6th ed; 1862.
- Blachar A, Federle MP, Brancatelli G, Peterson MS, Oliver 3rd JH, Li W. Radiologist performance in the diagnosis of internal hernia by using specific CT findings with emphasis on trans-mesenteric hernia. *Radiology* 2001;**221**:422–8.
- Leffall Jr LD, Quander J, Syphax B. Strangulation intestinal obstruction. *Archives of Surgery* 1965;**91**:592–6.
- Bizer LS, Liebling RW, Delany HM, Gliedman ML. Small bowel obstruction: the role of nonoperative treatment in simple intestinal obstruction and predictive criteria for strangulation obstruction. *Surgery* 1981;**89**:407–13.
- Passas V, Karavias D, Grilias D, Birbas A. Computed tomography of left paraduodenal hernia. *Journal of Computer Assisted Tomography* 1986;**10**:542–3.
- Meyers MA. Paraduodenal hernias. Radiologic and arteriographic diagnosis. *Radiology* 1970;**95**:29–37.
- Miller PA, Mezwa DG, Feczko PJ, Jafri ZH, Madrazo BL. Imaging of abdominal hernias. *Radiographics* 1995;**15**:333–47.
- Warshauer DM, Mauro MA. CT diagnosis of paraduodenal hernia. *Gastrointestinal Radiology* 1992;**17**:13–5.
- Day DL, Drake DG, Leonard AS, Letourneau JG. CT findings in left paraduodenal herniae. *Gastrointestinal Radiology* 1988;**13**:27–9.
- Harbin WP. Computed tomographic diagnosis of internal hernia. *Radiology* 1982;**143**:736.
- Bell-Thomson J, Vieta JO, Yiavasis AA. Paraduodenal hernias. *American Journal of Gastroenterology* 1977;**68**:254–9.
- Berardi RS. Paraduodenal hernias. *Surgery, Gynecology and Obstetrics* 1981;**152**:99–110.
- Dabas HT. *Gastrointestinal surgery, pathophysiology and management*, vol. 10; 2003. p. 365–6.
- Newsom BD, Kukora JS. Congenital and acquired internal hernias: unusual causes of small bowel obstruction. *American Journal of Surgery* 1986;**152**:279–85.
- Osadchy A, Keidar A, Zissin R. Small bowel obstruction due to a paracecal hernia: computerized tomography diagnosis. *Emergency Radiology* 2005;**11**:239–41.
- Patti R, Arcara M, Davi V, Di Marco V, Leo P, Di Vita G. Paraduodenal hernia: an uncommon cause of recurrent abdominal pain. *Giornale di Chirurgia* 2004;**25**:183–6.
- Tong RS, Sengupta S, Tjandra JJ. Left paraduodenal hernia: case report and review of the literature. *Australian and New Zealand Journal of Surgery* 2002;**72**:69–71.
- Davis R. Surgery of left paraduodenal hernia. *American Journal of Surgery* 1975;**129**:570–3.